

## General

### Title

Amyotrophic lateral sclerosis (ALS): percentage of patients diagnosed with ALS who were offered at least once annually assistance in planning for end of life issues.

### Source(s)

American Academy of Neurology (AAN). Amyotrophic lateral sclerosis performance measurement set. St. Paul (MN): American Academy of Neurology (AAN); 2012 Jul 31. 75 p.

## Measure Domain

### Primary Measure Domain

Clinical Quality Measures: Process

### Secondary Measure Domain

Does not apply to this measure

## Brief Abstract

### Description

This measure is used to assess the percentage of patients diagnosed with amyotrophic lateral sclerosis (ALS) who were offered at least once annually assistance in planning for end of life issues (e.g., advance directives, invasive ventilation, hospice).

### Rationale

Referral to palliative services varies considerably across different countries (Bede et al., 2011). End of life discussions will improve patient decision making with respect to disease management (Bede et al., 2011; Oliver et al., 2011; Albert et al., 1999; Mitsumoto et al., 2005; Nolan et al., 2008; Albert et al., 2005; Albert et al., 2009). Offering assistance in formulating an advanced care directive can initiate this discussion and re-discussion of the patient's preferences for life-sustaining treatments every 6 months (Albert et al., 1999; Mitsumoto et al., 2005; Nolan et al., 2008). Pain in amyotrophic lateral sclerosis (ALS) should be treated following accepted guidelines (Chiò et al., 2012; Handy et al., 2011;

Brettschneider et al., 2008). Physical therapy will aid in treating spasticity and pain (Morris et al., 2006). Discussion of respiratory support if the patient has dyspnea, other symptoms of hypoventilation or vital capacity (VC) less than 50% will allow patient to choose intervention or hospice (Morris et al., 2006; Tripodoro & De Vito, 2008). Early referral to hospice or home care teams well in advance of the terminal phase of ALS will facilitate the work of the hospice team and improve patient transition to hospice (McCluskey & Houseman, 2004; Krivickas, Shockley, & Mitsumoto, 1997). A medical social worker can help with financial issues. Medications for terminal dyspnea, pain and/or anxiety will improve quality of life (McCluskey & Houseman, 2004; Oliver et al., 2010; Dand & Sakel, 2010; Rabkin et al., 2005; Krivickas, Shockley, & Mitsumoto, 1997).

The following clinical recommendation statements are quoted verbatim from the referenced clinical guidelines and represent the evidence base for the measure:

Advance directives for palliative end-of-life care should be discussed early with the patient and carers, respecting the patient's social and cultural background (EFNS Task Force on Diagnosis and Management of Amyotrophic Lateral Sclerosis et al., 2012).

Offer assistance in formulating an advance care directive (Andersen et al., 2005).

Review the patients' wishes regarding their care and advance directives regularly (Heffernan et al., 2006).

Re-discuss the patient's preferences for life-sustaining treatments every 6 months (Andersen et al., 2005).

Initiate discussions on end-of-life issues whenever the patient asks or "opens the door" for end-of-life information and/or interventions (GPP) (Andersen et al., 2005). Treat pain in ALS following accepted guidelines (Andersen et al., 2005).

Initiate early referral to hospice or home care teams well in advance of the terminal phase of ALS to facilitate the work of the hospice team (Andersen et al., 2005).

Discuss options for respiratory support and end-of-life issues if the patient has dyspnea, other symptoms of hypoventilation or VC less than 50% (Andersen et al., 2005).

Treat terminal dyspnea and/or pain with opioids alone or in combination with benzodiazepines if anxiety is present (Andersen et al., 2005).

## Evidence for Rationale

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## Primary Health Components

Amyotrophic lateral sclerosis (ALS); advance directives; invasive ventilation; hospice

## Denominator Description

All patients with a diagnosis of amyotrophic lateral sclerosis (ALS) (see the related "Denominator Inclusions/Exclusions" field)

## Numerator Description

Patients who were offered at least once annually assistance in planning for end of life issues (e.g., advance directives, invasive ventilation, or hospice)

## Evidence Supporting the Measure

### Type of Evidence Supporting the Criterion of Quality for the Measure

A clinical practice guideline or other peer-reviewed synthesis of the clinical research evidence

A formal consensus procedure, involving experts in relevant clinical, methodological, public health and organizational sciences

A systematic review of the clinical research literature (e.g., Cochrane Review)

One or more research studies published in a National Library of Medicine (NLM) indexed, peer-reviewed journal

## Additional Information Supporting Need for the Measure

Importance of Topic

### *Prevalence and Incidence*

Amyotrophic lateral sclerosis (ALS), also known as Lou Gehrig's disease, is a type of motor neuron disease that is a rapidly progressive and fatal neurological disease (National Institute of Neurological Disorders and Stroke [NINDS], 2013).

Twenty thousand to 30,000 people in the United States (U.S.) have ALS (NINDS, 2013).

Five thousand people are diagnosed with ALS in the U.S. annually (NINDS, 2013).

ALS is one of the most common neuromuscular diseases worldwide (NINDS, 2013).

In 90% to 95% of all ALS cases the disease occurs apparently at random with no clearly associated risk factors (NINDS, 2013).

Five percent to 10% of all ALS cases are inherited (NINDS, 2013).

Twenty percent of all familial cases result from a specific genetic defect that leads to mutation of the enzyme known as superoxide dismutase 1 (SOD1) (NINDS, 2013).

No cure exists for ALS. Newer pharmacotherapy agents have been found to reduce the progression, but not halt the disease development (NINDS, 2013).

The prevalence of ALS is said to be between six and eight cases per 100,000 in the population. Using the higher prevalence estimate and data from the 2000 U.S. census, nearly 22,600 Americans are living with ALS at any one time. Since ALS is a disease of aging, as the U.S. population increases and ages, an increase in the prevalence of ALS can be anticipated (ALS Association, 2012). Cognitive dysfunction is seen in 20% to 50%, while only 3% to 5% develop dementia that is usually of frontotemporal type (Strong et al., 2009). Consensus criteria for diagnosis have recently been reported (Strong et al., 2009). Death due to respiratory failure follows on average 2 to 4 years after onset, but a small group may survive for a decade or more (Haverkamp, Appel, & Appel, 1995). The mean age of onset is 47 to 52 years in familial cases (FALS) and 58 to 63 years in sporadic (SALS) cases (Bobowick & Brody, 1973). The lifetime risk for developing ALS for individuals aged 18 years has been estimated to be 1 in 350 for men and 1 in 420 for women (Armon, 2007) with male sex, increasing age and hereditary disposition being the main risk factors (Heffernan et al., 2006).

### *Mortality and Morbidity*

Most patients with ALS die within 2 to 5 years of onset (Lechtzin et al., 2002). Only 10% of ALS patients survive for 10 years or more (Miller et al., "Drug, nutritional," 2009). Treatment of respiratory insufficiency improves survival, quality of life and respiratory symptoms (Lechtzin et al., 2002; Miller et al., "Drug, nutritional," 2009). The diagnosis and management of respiratory insufficiency is critical because most deaths from ALS are due to respiratory failure (Lechtzin et al., 2002; Miller et al., "Drug, nutritional," 2009; EFNS Task Force on Diagnosis and Management of Amyotrophic Lateral Sclerosis et al., 2012; Laird et al., 2001). Falls surveillance will lead to interventions to prevent falls and decrease fall related deaths in ALS patients. Falls are an independent predictor of adverse health outcomes (Gil et al., 2008). Fall related deaths occur in 1.7% of ALS patients (Rubenstein & Josephson, 2002). Several specific risk factors for falls have been identified, including muscle weakness, deficits in gait or balance, visual deficits, arthritis, impairments in activities of daily living, depression, and cognitive impairment (Ringholz et al., 2005). Studies confirm the presence of cognitive impairment in 50% of patients with ALS and particularly implicate executive dysfunction and mild memory decline in the disease process (Laird et al., 2001). More severe impairment occurs in a subset of patients with ALS and has features consistent with frontal temporal dementia (FTD) (Phukan, Pender, & Hardiman, 2007; Gordon et al., 2007). Recent studies have demonstrated the feasibility of screening patients in a busy specialized ALS clinic (Flaherty-Craig et al., 2009; Woolley & Katz, 2011), but this is still not routinely practiced. A fuller characterization of the extent of cognitive and behavioral dysfunction in ALS has important implications given that it shortens survival (Elamin et al., 2011), and the burden and stress for carers of patients with FTD is very great. It also has relevance to effective communication, legal issues and end-of-life decision making by patients with motor neuron disease (MND) (Elamin et al., 2011). Pseudobulbar affect (PBA), excessive laughing or crying, or involuntary emotional expression disorder affects 20% to 50% of patients with ALS, especially in pseudobulbar palsy (McCullagh et al., 1999). Patients are embarrassed and isolated by these symptoms, which in turn greatly diminishes the patients' quality of life. Sialorrhea, or drooling, is embarrassing, socially isolating, and is associated with aspiration pneumonia. The prevalence is estimated at 50%, and 70% of patients receiving oral medications for treatment reported benefit (Laird et al., 2001; Miller et al., "Multi-disciplinary," 2009). Fatigue may be a symptom of depression, poor sleep, abnormal muscle activation, immobility, or respiratory dysfunction. Fatigue diminishes quality of life for patients with ALS. Fatigue was a side effect of therapy in 26% of patients taking riluzole vs. 13% taking placebo (Bensimon, Lacomblez, & Meininger, 1994). Asthenia occurred in 18% of patients taking riluzole vs. 12% of patients taking placebo in a larger study (Lacomblez et al., 1996). The prevalence of depression in ALS ranges from 0% to 44%, although systematic studies suggest 10% in advanced ALS (Laird et al., 2001; Wicks et al., 2007). Depression shortens survival and

lowers quality of life for patients with ALS (Phukan, Pender, & Hardiman, 2007). There is consensus among experts that depression should be treated in patients with ALS (Laird et al., 2001); however, there are no controlled studies of benefit or harm.

Insomnia is common in ALS and may be a symptom of early respiratory weakness, underlying anxiety, depression, or pain (Hetta & Jansson, 1997). There is a concern that sedative/hypnotic agents may suppress the respiratory drive in patients with ALS.

Weight loss is a key prognostic indicator for ALS with the risk of death increased 7-fold when body mass index is less than 18.5 kg/m<sup>2</sup> (Marin et al., 2011; Lehericey et al., 2012; Spataro et al., 2011; Desport et al., 1999; Vaisman et al., 2009; Dupuis et al., 2008).

ALS patients have dysarthria in nearly all bulbar onset patients and nearly 40% of ALS patients with spinal onset. More than 95% of ALS patients cannot speak before death and patients who accept gastrostomy tube, non-invasive ventilation or tracheostomy-ventilation have a greater need for augmentative alternative communication as the disease progresses (Ball, Beukelman, & Pattee, "Communication," 2004; Ball, Beukelman, & Pattee, "Acceptance," 2004; Mathy, Yorkston, & Gutmann, 2000; Beukelman, Fager, & Nordness, 2011).

End of life discussions will improve patient decision making with respect to disease management (NINDS, 2013; ALS Association, 2012; Strong et al., 2009; Haverkamp, Appel, & Appel, 1995; Bobowick & Brody, 1973; Heffernan et al., 2006). Pain in ALS should be treated following accepted guidelines (Oliver et al., 2011; Albert et al., 1999, Mitsumoto et al., 2005; Nolan et al., 2008; Albert et al., 2005; Albert et al., 2009).

### *Office Visits and Hospital Stays*

One study's significant findings were that common morbidities increased over time (pneumonia [38.1% to 47.3%], respiratory failure [26.9% to 35.5%], and nutritional deficiency [43.0% to 56.3%]); the median length of stay dropped from 6 to 4 days; mean hospital charges increased from \$21,574 to \$24,314; the proportion of hospital deaths decreased over time (17.6% to 14.6%), whereas the proportion discharged to home health/hospice care (14.0% to 18.2%) and to long-term care facilities (13.2% to 27.9%) increased. The odds ratio (OR) of death was 5.03 (95% CI: 4.57 to 5.54) for those admitted with respiratory failure, 1.36 (1.24 to 1.50) for those with pneumonia, and 0.84 (0.77 to 0.92) for those with nutritional deficiency. The high OR of death in patients admitted for pneumonia or respiratory failure is likely associated with more advanced disease, whereas the protective effect of admission for nutritional deficiency is consistent with the predominance of bulbar symptoms and admission earlier in the disease. The trends during the 15 years of this administrative data set were for increasing comorbidities and higher utilization of end-of-life care (Dubinsky, Chen, & Lai, 2006).

### *Family Caregiving*

Caregiver burden was correlated to their level of depression and quality of life and, differently from other chronic disorders, increased with the worsening of patients' disability. ALS patients have a good objective perception of their impact on caregivers (Chiò et al., 2005).

Recent studies assessing caregivers' burden in chronic neurologic disorders have found some features shared by caregivers: the perceived burden exceeds the objective measures of patients' impairment, the amount of burden is independent of diagnosis, and the patients' cognitive functioning is an important factor in determining the level of burden (Thommessen et al., 2002).

### *Cost*

ALS is a difficult to diagnose, fatal, progressive degenerative disease with an average survival time of 2 to 5 years. Percutaneous endoscopic gastrostomy (PEG) and bi-level intermittent positive pressure (BIPAP) ventilation may be the major interventions leading to longer survival of patients with ALS. Riluzole has been shown to have modest effects on survival (as opposed to functional) gains and is currently the only drug approved for the treatment of ALS. Mechanical ventilation (via a tracheostomy tube) is expensive, but is widely used in later stage patients with ALS in the U.S. A review of nine cost-effectiveness studies of riluzole found the following: drug costs and survival



gains are the major drivers of cost effectiveness; survival gains are estimated from truncated databases with a high degree of uncertainty; more accurate stage-specific utility weights based on patients who agreed to treatment are needed; case incidence-based evaluations should be carried out; cost-effectiveness ratios are insensitive to discount rates; employment and caregiver issues or externalities have been widely ignored; threshold acceptance cost-effectiveness values are ill-defined and evaluations are not generalizable to other countries because of cost and treatment style differences. On account of the high degree of uncertainty pertaining to survival gains and the relatively high costs per life years or quality-adjusted life-years gained, and while acknowledging that not every therapy has to be cost effective (e.g., orphan drugs), it is still inconclusive as to whether or not riluzole can be considered as cost-effective therapy for ALS (Ginsberg & Lowe, 2002).

## Disparities

All races and ethnic backgrounds are affected by ALS (NINDS, 2013).

ALS most common in individuals 40 to 60 years old, but younger and older people can develop the disease (NINDS, 2013).

Men are more likely to develop ALS than women. Studies suggest an overall ratio of about 1.5 men to every woman who develops ALS in Western countries (ALS Association, 2012).

## Opportunity for Improvement

Palliative care should be adopted from the time of diagnosis (Bede et al., 2011). Many patients are not adequately informed about advance directives and end of life decision making and many hospice workers are not familiar with ALS (Mandler et al., 2001; Miller et al., "Outcomes," 2009; Bradley et al., 2004). Management of the terminal phase of ALS is unsatisfactory in 33% to 61% of cases in Europe (Borasio et al., 2001) and only 8% of palliative care units are involved from the time of diagnosis (Oliver & Webb, 2000). The current system of palliative care in the United States is highly decentralized (National Hospice and Palliative Care Organisation [NHPCO], 2009). Between 60% and 88% of patients die in a medical facility in some countries and not at home, while over 58% of seriously ill ALS patients do not have hospice care (Spataro et al., 2010; Gil et al., 2008; Krivickas, Shockley, & Mitsumoto, 1997). Approaches to end of life care vary widely and are not standardized either in timing or content (Bede et al., 2011; Elman et al., 2006).

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## Extent of Measure Testing

This measure is being made available without any prior testing. The American Academy of Neurology (AAN) recognizes the importance of testing of all of its measures and encourages testing of the amyotrophic lateral sclerosis (ALS) measurement set for feasibility and reliability by organizations or individuals positioned to do so. The AAN welcomes the opportunity to promote the initial testing of these measures and to ensure that any results available from testing are used to refine the measures before implementation.

## Evidence for Extent of Measure Testing

American Academy of Neurology (AAN). Amyotrophic lateral sclerosis performance measurement set. St. Paul (MN): American Academy of Neurology (AAN); 2012 Jul 31. 75 p.

## State of Use of the Measure

### State of Use

Current routine use

## Current Use

not defined yet

## Application of the Measure in its Current Use

### Measurement Setting

Ambulatory/Office-based Care

Home Care

Hospital Outpatient

Skilled Nursing Facilities/Nursing Homes

### Professionals Involved in Delivery of Health Services

not defined yet

### Least Aggregated Level of Services Delivery Addressed

Individual Clinicians or Public Health Professionals

### Statement of Acceptable Minimum Sample Size

Does not apply to this measure

### Target Population Age

Unspecified

### Target Population Gender

Either male or female

## National Strategy for Quality Improvement in Health Care

### National Quality Strategy Aim

Better Care

### National Quality Strategy Priority

Person- and Family-centered Care

Prevention and Treatment of Leading Causes of Mortality

# Institute of Medicine (IOM) National Health Care Quality Report Categories

## IOM Care Need

End of Life Care

Living with Illness

## IOM Domain

Effectiveness

Patient-centeredness

## Data Collection for the Measure

### Case Finding Period

Unspecified

### Denominator Sampling Frame

Patients associated with provider

### Denominator (Index) Event or Characteristic

Clinical Condition

### Denominator Time Window

not defined yet

### Denominator Inclusions/Exclusions

#### Inclusions

All patients with a diagnosis of amyotrophic lateral sclerosis (ALS)

Note: Refer to the original measure documentation for International Classification of Diseases, Ninth Revision, Clinical Modification (ICD-9-CM) diagnosis codes and Current Procedural Terminology (CPT) Evaluation and Management (E/M) service codes.

#### Exclusions

Documentation of a medical reason for not offering at least once annually assistance in planning for end of life issues (e.g., patient in hospice and already in terminal phase)

### Exclusions/Exceptions

not defined yet

## Numerator Inclusions/Exclusions

### Inclusions

Patients who were offered at least once annually assistance in planning for end of life issues (e.g., advance directives, invasive ventilation, or hospice)

### Exclusions

Unspecified

## Numerator Search Strategy

Fixed time period or point in time

## Data Source

Administrative clinical data

Electronic health/medical record

Paper medical record

## Type of Health State

Does not apply to this measure

## Instruments Used and/or Associated with the Measure

Unspecified

## Computation of the Measure

### Measure Specifies Disaggregation

Does not apply to this measure

## Scoring

Rate/Proportion

## Interpretation of Score

Desired value is a higher score

## Allowance for Patient or Population Factors

not defined yet

## Standard of Comparison



not defined yet

# Identifying Information

## Original Title

Measure #10: ALS end of life planning assistance.

## Measure Collection Name

Amyotrophic Lateral Sclerosis Performance Measurement Set

## Submitter

American Academy of Neurology - Medical Specialty Society

## Developer

American Academy of Neurology - Medical Specialty Society

## Funding Source(s)

Unspecified

## Composition of the Group that Developed the Measure

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*Muscular Dystrophy Association:* Valerie Cwik, MD

*Patient Representative:* Christine Jasch, OTR/L

*Insurance Representatives:* Fredrik Tolin, MD, MBA (Humana)

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## Financial Disclosures/Other Potential Conflicts of Interest

Unspecified

## Adaptation

This measure was not adapted from another source.

## Date of Most Current Version in NQMC

2012 Jul

## Measure Maintenance

Unspecified

## Date of Next Anticipated Revision

Unspecified

## Measure Status

This is the current release of the measure.

## Measure Availability

Source available from the [American Academy of Neurology \(AAN\) Web site](#) .

For more information, contact AAN at 201 Chicago Avenue, Minneapolis, MN 55415; Phone: 800-879-1960; Fax: 612-454-2746; Web site: [www.aan.com](http://www.aan.com) .

## NQMC Status

This NQMC summary was completed by ECRI Institute on March 8, 2016. The information was not verified by the measure developer.

## Copyright Statement

This NQMC summary is based on the original measure, which is subject to the measure developer's copyright restrictions.

## Production

Source(s)

American Academy of Neurology (AAN). Amyotrophic lateral sclerosis performance measurement set. St. Paul (MN): American Academy of Neurology (AAN); 2012 Jul 31. 75 p.

## Disclaimer

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